Surgical Options for Discrete Supravalvar Aortic Stenosis

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Supravalvar aortic stenosis is a rare anomaly in which there is an exaggerated narrowing at the junction of the sinuses of Valsalva with the ascending aorta, ie, at the sinotubular junction. This anomaly is often part of Williams syndrome and may be associated with generalized hypoplasia of the ascending aorta and more distal arterial tree as well as with stenoses in the pulmonary artery tree. A recent analysis has demonstrated that even in children who have a relatively severe form of supravalvar aortic and pulmonic stenosis the longterm outlook can be quite satisfactory so long as an aggressive approach employing both surgical and interventional catheter procedures is undertaken.[1]

Both McGoon and Starr and associates independently reported a series of patch enlargements for localized supravalvar aortic stenosis in 1961.[2] A major conceptual and therapeutic advance was made in 1976 when Doty and colleagues described the extended aortoplasty technique for this lesion.[3]

INDICATIONS FOR SURGERY

Supravalvar aortic stenosis is probably a progressive lesion in the majority of children and therefore should be treated before left ventricular hypertrophy has become severe. Early treatment will also decrease the risk of acute cardiac arrest and damage to the aortic valve. It may also decrease the probability of progressive coronary ostial stenosis.

In general a Doppler derived peak gradient of more than 40 to 50 mm Hg in association with definite evidence by 2-D echo imaging of an important narrowing of the sinotubular junction should be an indication to proceed to surgery. If the gradient is less than 30–40 mmHg and there is no evidence of left ventricular hypertrophy it is reasonable to follow the child with regular, eg, six monthly, echocardiographic evaluations.

If the child has evidence of Williams syndrome and the echocardiogram suggests that the lumen of the ascending aorta is small distally with stenoses extending into the arch vessels then imaging with MRI or cardiac catheterization should be undertaken to determine the extent of reconstruction that will be required. Preoperative assessment should also carefully exclude the presence of associated supravalvar pulmonary stenosis or mediastinal branch or peripheral pulmonary artery stenoses.

TECHNICAL CONSIDERATIONS

A careful assessment must be made preoperatively as to whether the repair should be limited to the area of the sinotubular junction or if a more extensive reconstruction of the ascending aorta and aortic arch is required. If there is obvious narrowing of the ascending aorta it is generally best to err on the side of extending a patch across the undersurface of the aortic arch. Failure to do this will simply transfer the supravalvar gradient to the distal ascending aorta. By extending a patch at least beyond the takeoff of the arch vessels the cardiac output can be decompressed into the arch vessels though there may still be some residual gradient at the toe of the patch.

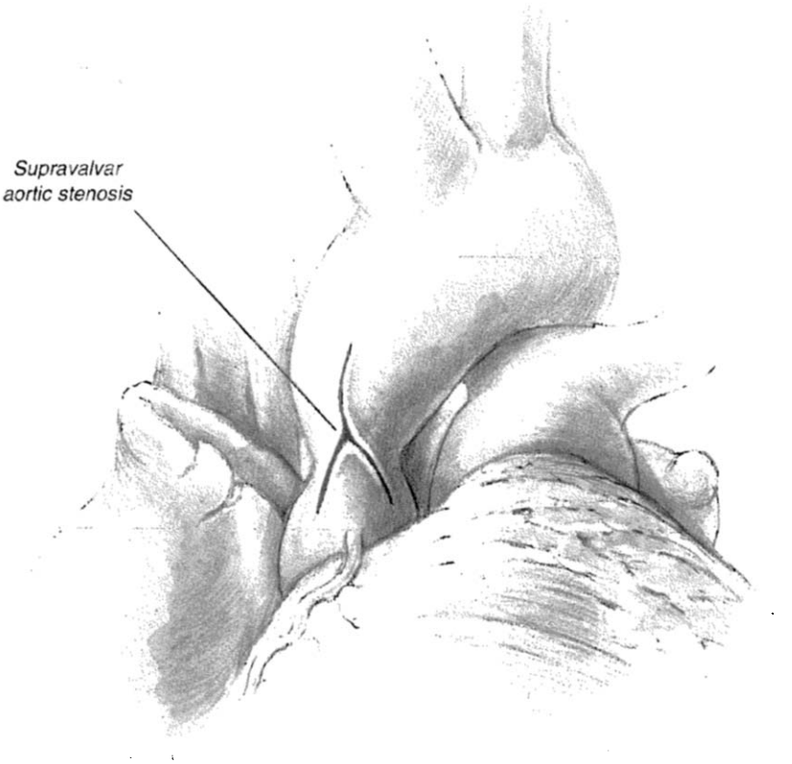
MANAGEMENT OF DISCRETE SUPRAVALVAR AORTIC STENOSIS

Single Patch

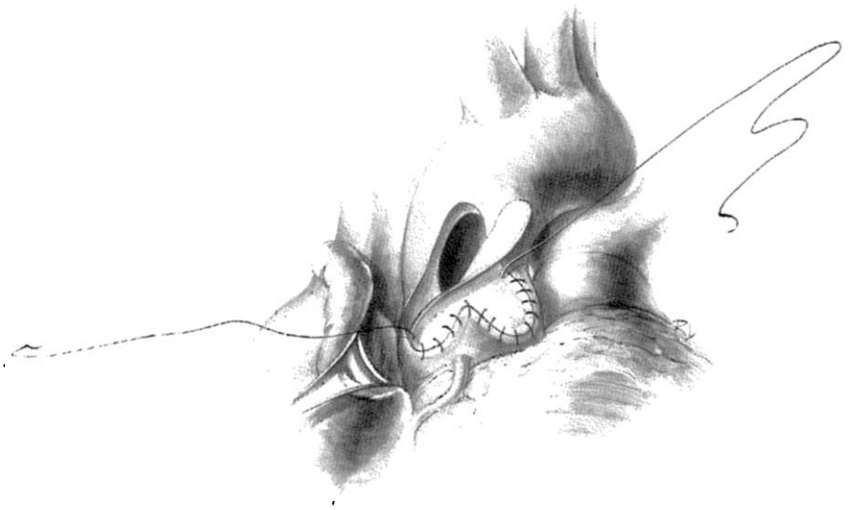
A recent review of the experience at Children's Hospital by Stamm and coworkers[1] has demonstrated that a single patch technique in the noncoronary sinus results in a significantly inferior outcome relative to a bifurcated patch or symmetric three patch technique. Therefore, we limit use of a single patch extending into the noncoronary sinus to those instances where relief of mild to moderate supravalvar stenosis is being undertaken in conjunction with relief of left ventricular outflow tract obstruction at other levels.

A. Inverted Bifurcated Patch Technique

This technique was originally described by Doty[3] and is appropriate for moderate or moderate to severe supravalvar aortic stenosis that does not involve important narrowing of the left coronary sinus of Valsalva.



A1 With ascending aortic cannulation and a single venous cannula in the right atrium and following the application of the aortic cross clamp and infusion of cardioplegia solution, a longitudinal incision is made on the anterior surface of the proximal ascending aorta. The incision is bifurcated into the middle of thenoncoronary sinus as well as into the right coronary sinus to the left of the right coronary ostium passing through the thickened sinotubular ridge. It is important that the right coronary ostium be carefully visualized and that the incision has adequate clearance from the right coronary ostium to allow subsequent suturing. Following completion of the bifurcated incision the right coronary ostium sits on a small triangle of tissue directly anteriorly.



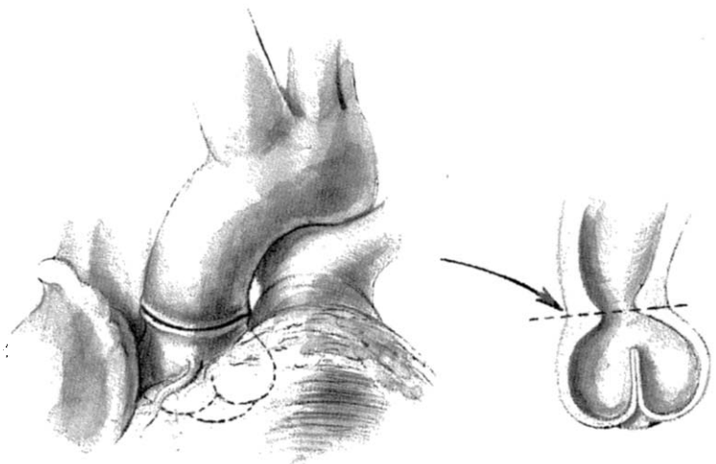
A2 A generous pantaloon shaped patch is now sutured into the two sinuses of Valsalva. It is important to understand that the goal is to create bulging sinuses of Valsalva similar to those seen normally so that the patch should appear quite a bit larger than one would initially anticipate. Interestingly despite placing very generous patches in the two anterior sinuses it is rare that sufficient distortion of the aortic valve is created that aortic regurgitation ensues.

The choice of patch material is dependent on the age of the patient. In the smaller, younger patient it is preferable to use autologous pericardium treated with 0.6% glutaraldehyde for 20–30 minutes. Pericardium is very much more hemostatic than synthetic alternatives. In the larger older patient it is probably wise to use collagen impregnated crimped Dacron (eg, Hemashield). It is not wise to use PTFE (eg, Goretex) because of excessive needlehole bleeding in this location even when PTFE suture is employed.

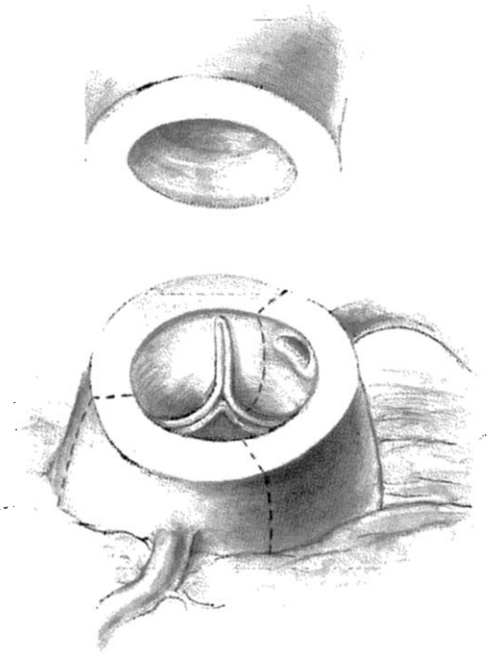
If this procedure is undertaken in conjunction with patching of the ascending aorta and arch one patch started in the arch and distal ascending aorta is used. The usual maneuvers are undertaken for de-airing the left heart including allowing an aortic vent site to bleed freely at the time of release of the aortic cross clamp.

B. Symmetric Three Patch Approach

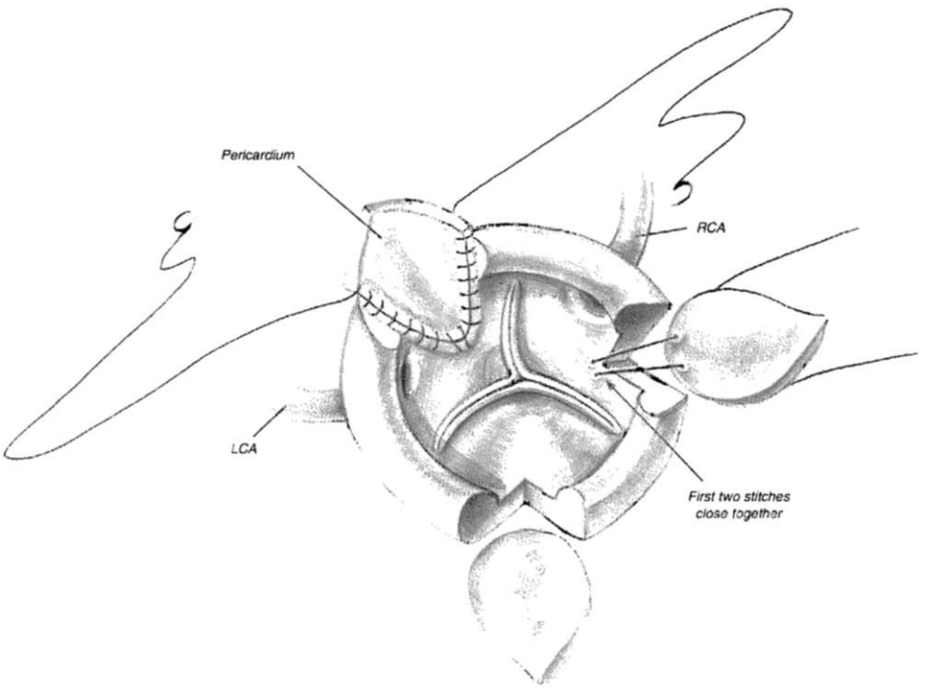
If there is important narrowing of the left coronary sinus as is often seen with severe forms of supravalvar stenosis, one option is to place three independent patches, one in each sinus of Valsalva. This can be achieved by advancement of the ascending aorta (see below). An alternative is to use autologous pericardium.



B1 The ascending aorta is divided transversely at the level of the sinotubular junction or slightly above.



B2 Incisions are carried down into each of the sinuses of Valsalva. In the case of the left coronary sinus the incision is just to the right of the left coronary ostium. In the case of the right coronary sinus the incision is just to the left of the right coronary ostium. The incision is carried well into the sinus of Valsalva with care to avoid injury to the valve leaflets.



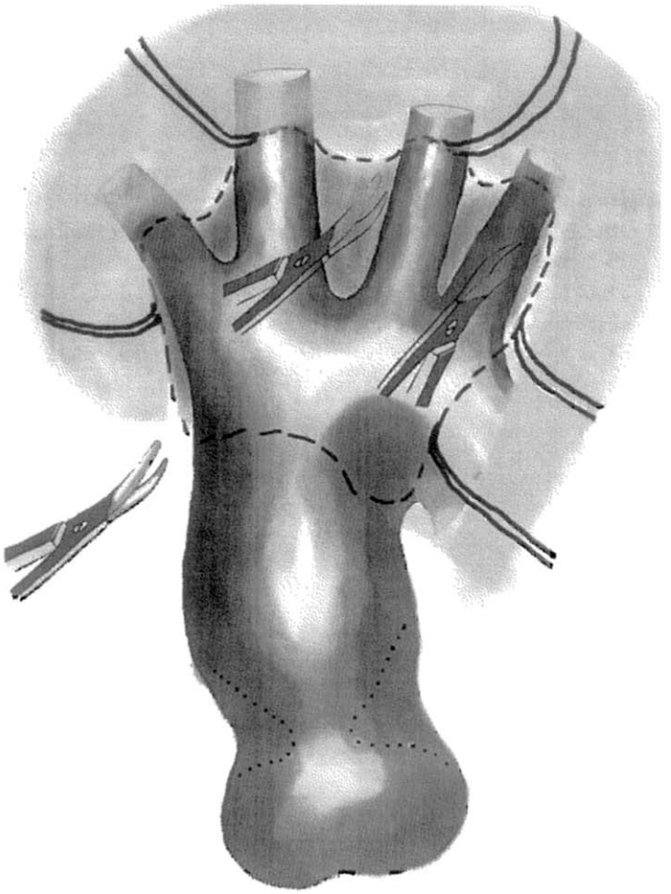
B3 Teardrop shaped patches of autologous pericardium treated with 0.6% glutaraldehyde for 30 minutes are sutured into each of the sinuses of Valsalva. It is important to use a differential suturing technique, particularly at the apex of the incision. Bites are widely spaced on the pericardium and closely spaced in the sinus of Valsalva. This differential spacing allows optimal supplementation of the sinus of Valsalva. As with the pantaloon shaped patch the goal should be to achieve a bulging normal shaped sinus of Valsalva so the pericardial patch is relatively large and redundant.

It is very common to extend the two anterior patches into the ascending aorta. A longitudinal incision is made on the anterior ascending aorta. The two anterior patches are sutured together along their contiguous margins and extend into the distal aortotomy. Posteriorly the aorta is reconstituted directly to the supplemented sinotubular junction.

C. Direct Anastomosis Technique

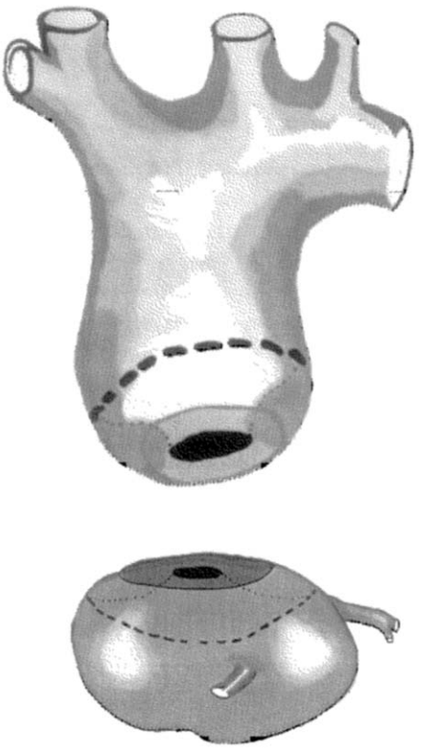
The technique of direct anastomosis can be applied when supravalvar aortic stenosis is well circumscribed and limited to the area in and around the sinotubular junction.

Aortic Mobilization



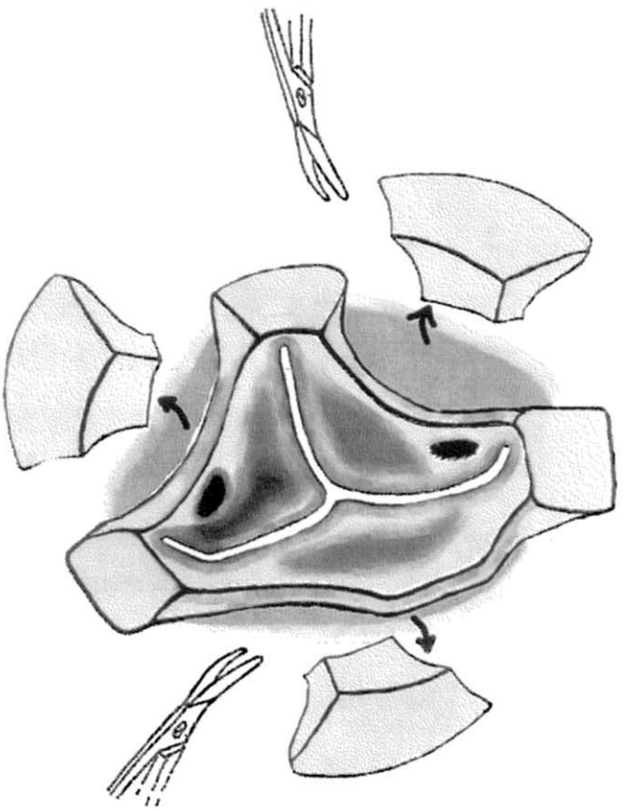
C1 Mobilization of the distal ascending aorta and arch branches allows sufficient mobility for resection of discrete supravalvar aortic stenosis with direct anastomosis. For those cases with more extensive involvement of the ascending aorta and aortic arch these techniques can still be combined with patching techniques to achieve lasting relief of obstruction while preserving aortic valve function and growth.

Lines of Resection



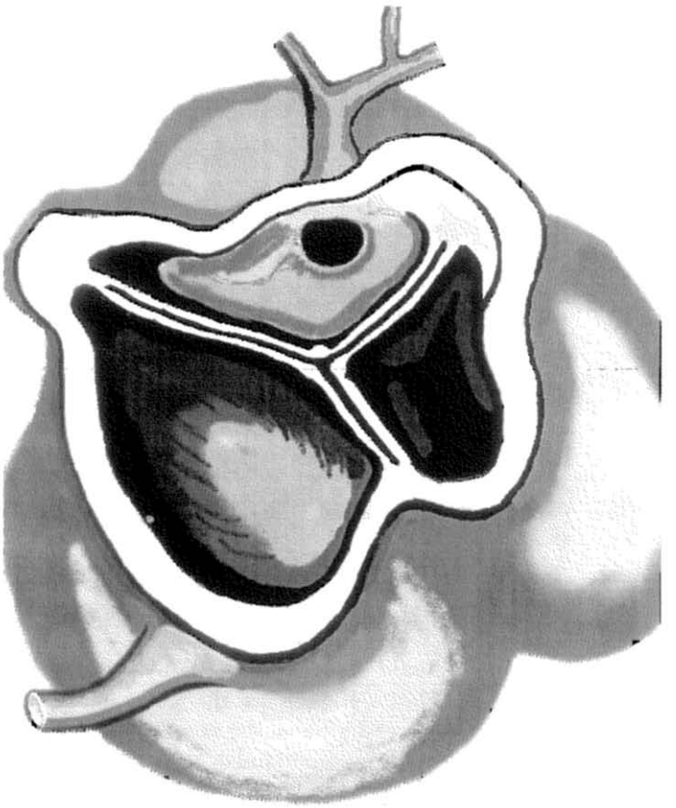
C2 Complete resection of the thickened aortic wall is essential for adequate relief and involves resection of this tissue from proximal and distal stumps after division of the aorta just distal to the aortic valve commissural peaks.

Proximal Resection 1

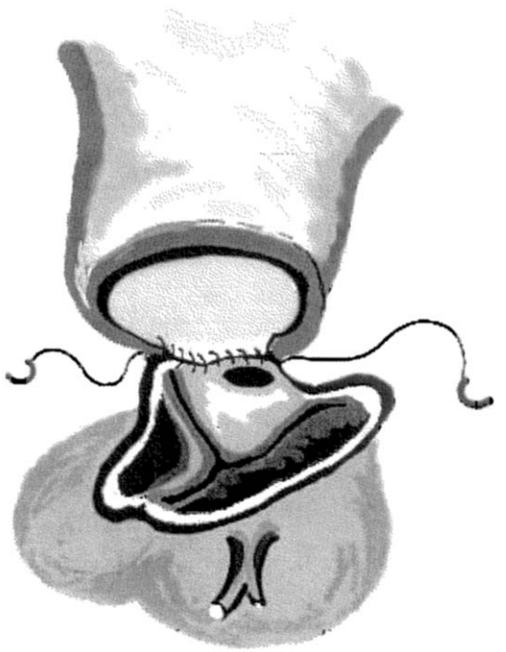


C3 Resection lines must extend into the aortic sinuses and closely skirt the coronary orifices. This is most safely achieved by resecting this tissue in the noncoronary sinus first to allow the proximal stump to open sufficiently to view the other sinuses accurately.

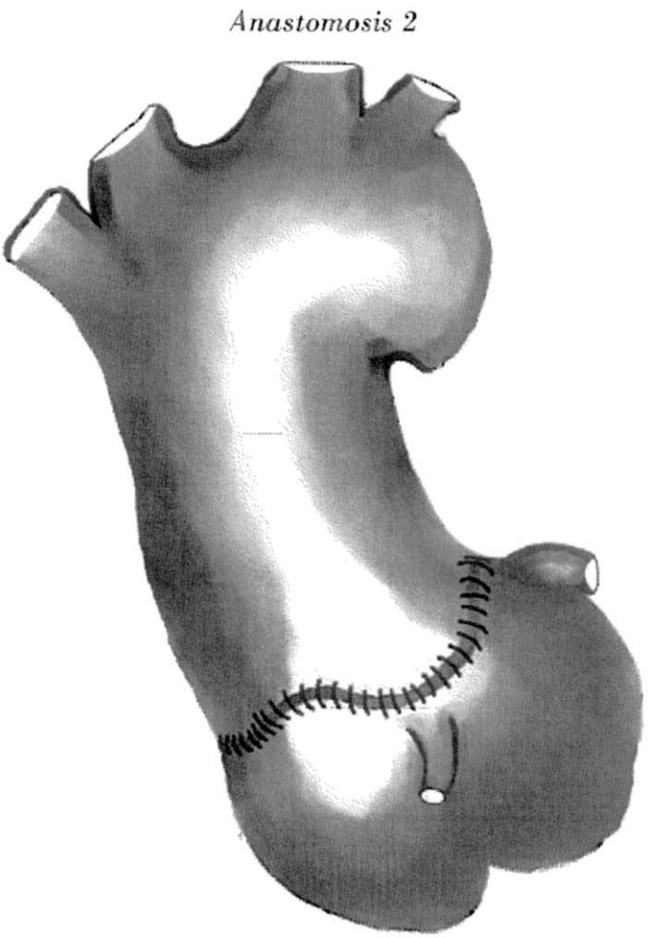
Proximal Resection 2



C4 The proximal aortic stump opens very well after adequate resection. Note the proximity of the coronary orifices to the intended reconstruction suture line.



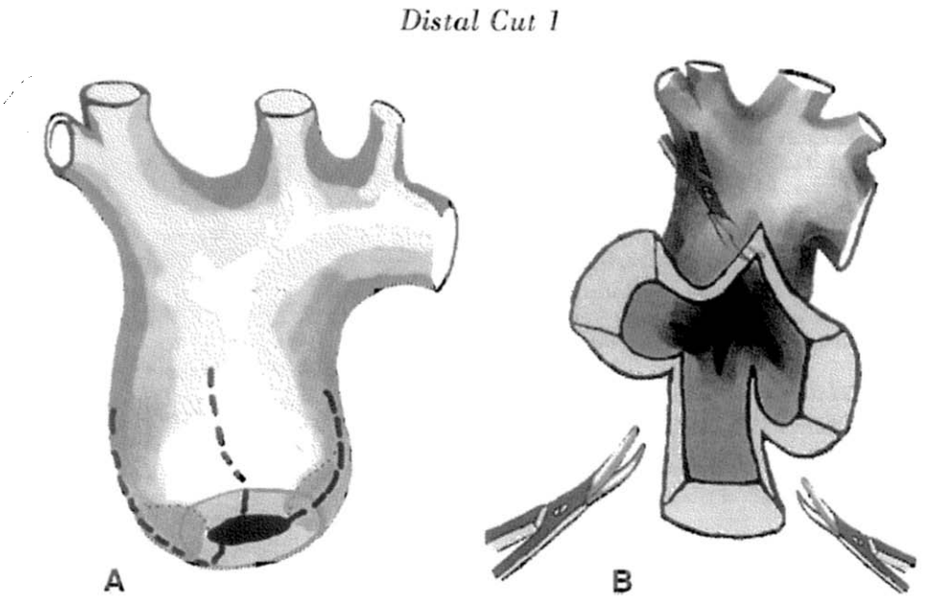
C5 Reconstruction should start near the left coronary orifice so that this can be well seen as the anastomosis proceeds. Sutures will of necessity be on the verge of the left coronary orifice. The suture line should finish away from the right coronary orifice for the same reason.



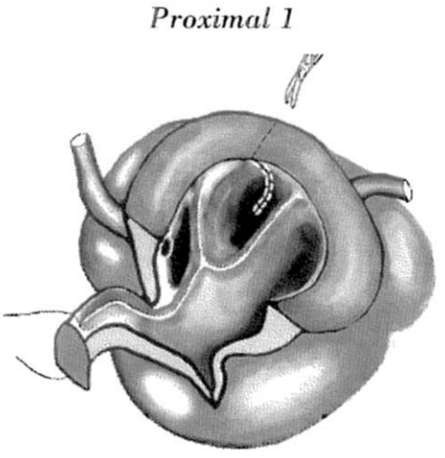
C6 The mobilization of the arch branches allows approximation without tension and tissue resection allows a normal sized reconstructed sino tubular junction with sustained relief of supra valvar aortic stenosis.

D. Modified Direct Anastomosis Technique

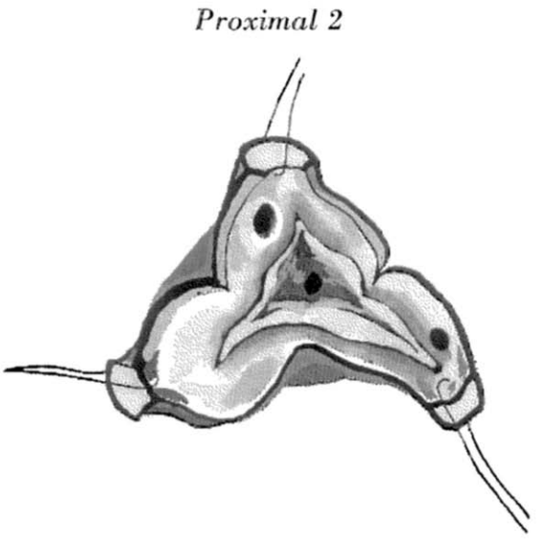
This technique also employs autologous aortic reconstruction of the sinuses of Valsalva. Rather than direct anastomosis, however, the ascending aorta is spatulated in such a fashion as to interdigitate with the incisions in the three sinuses of Valsalva as originally described by Myers and coworkers.[4]



D1 The three lines of incision should extend well into the distal ascending aorta and correspond to the length of the incisions into the aortic sinuses and be positioned so that they are 120° to these sinus incisions. This allows the three tongues of tissue cut to advance into the corresponding sinuses.

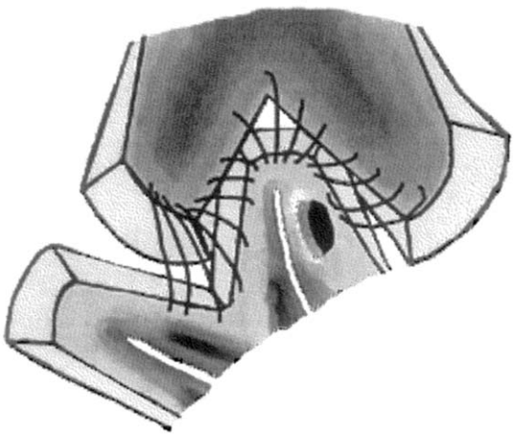


D2 The first incision into the proximal stump should be into the noncoronary sinus so that subsequent cuts can more fully appreciate the positioning of the left and right coronary arteries within their respective sinuses.



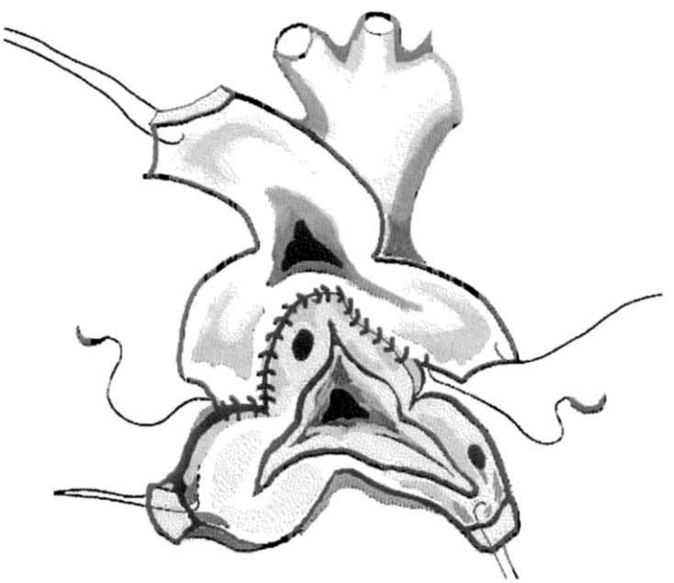
D3 The proximal stump opens completely after these incisions and the normal nature of the aortic valve leaflets is easily appreciated. The closeness of the coronary orifices to the lines of incision is often exquisite.

Near and Far Suturing



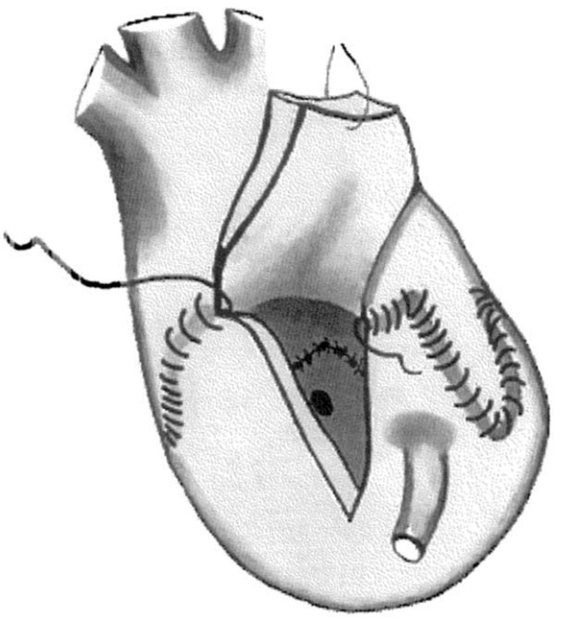
D4 The three tongues of tissue that advance into the proximal incisions require careful near and far suturing to maintain the accuracy of the three tongues positioning around the circumference to the proximal stump.

Anastomosis 1



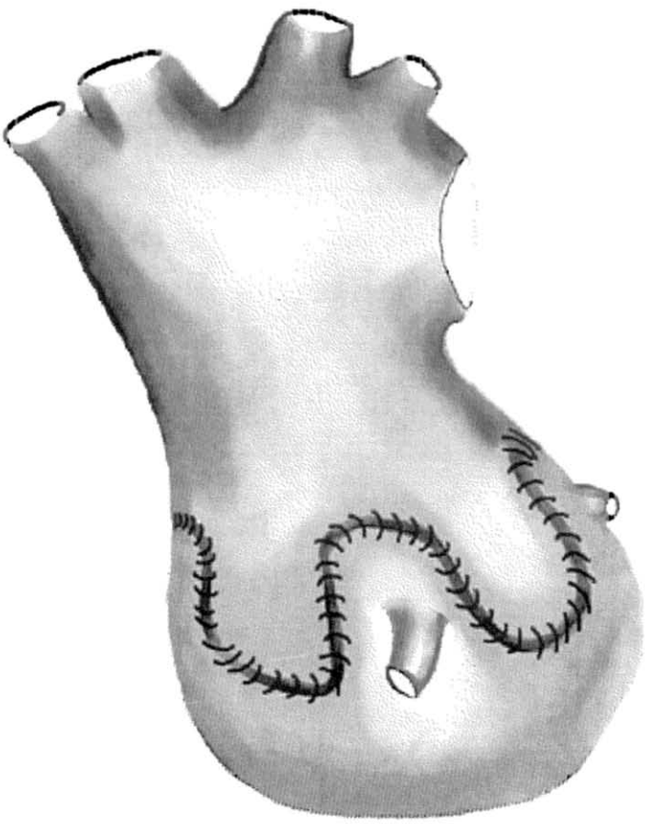
D5 Joining of the aorta should begin posteriorly and proceed to completion anteriorly away from the right coronary orifice. The thickened nature of the tissue is no impediment to an accurate suture line.

Anastomosis 2



D6 Tissue advancement into the sinuses is very adequate.

Anastomosis 3



D7 The completed long suture line successfully enlarges the sino tubular junction without tissue resection and provides sustained relief of supra valvar aortic stenosis with preservation of aortic growth and normal valve function.

RESULTS

A recent review described a 41 year experience with 75 patients undergoing operations to treat congenital supravalvar aortic stenosis at Children's Hospital Boston up to 1998.[J] In 34 patients single patch enlargement of the noncoronary sinus was undertaken (almost all early in the series). Other procedures included an inverted bifurcated patch plasty in 35 patients and three-sinus reconstruction of the aortic root in 6 patients. There were seven early deaths. Among early survivors 100% were alive at 5 years, 96% were alive at 10 years and 77% were alive at 20 years. Diffuse stenosis of the ascending aorta was a risk factor for both survival and reoperation ($P < 0.01$ for each). Patients with multiple sinus reconstructions had a significantly lower probability of reoperation. Residual gradients were also lower after multiple sinus reconstruction of the aortic root as was the prevalence of moderate aortic regurgitation at follow-up. The authors concluded from this study that results of surgery for supravalvar aortic stenosis improved greatly after the introduction of more symmetric reconstruction of the aortic root. Multiple sinus reconstruction also resulted in superior hemodynamics and was associated with reductions in both mortality rate and the need for reparation.

In 2001 Kang, Nunn, Andrews and Chard[5] updated the results of their experience with direct anastomosis repair of supravalvar aortic stenosis.[6] One patient who had had a preoperative gradient of 120 mm and who underwent surgery at 6 months of age required subsequent surgery for subaortic stenosis. There was no residual gradient at the level of the supravalvar area. Another patient who underwent surgery at 2.5 years of age for a gradient of 100 mm has a residual gradient of 25 mm at age 7.5 years. The authors continue to believe that repair by direct end to end anastomosis is a useful technique in selected cases of discrete supravalvar aortic stenosis.

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